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Mehdi Fathi^a, Marjan Joudi^b, Yaser Rajabnejad^c, Azra Izanloo^d, Mona Joudi^{e,*}, Mehran Hiradfar^f, Alireza Sabzevari^a

^a Department of Anesthesiology, Surgical Oncology Research Center, Imam Reza Hospital, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

^b Department of Surgery, Surgical Oncology Research Center, Imam Reza Hospital, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

^c Department of General Surgery, Surgical Oncology Research Center, Imam Reza Hospital, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

^d Department of Radiology, Faculty of Paramedical Sciences, Mashhad University of Medical Sciences, Mashhad, Iran

e Department of Radiation Oncology, Omid Hospital, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

^f Department of Pediatric Surgery, Surgical Oncology Research Center, Mashhad University of Medical Sciences, Mashhad, Iran

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ABSTRACT

A lack of consensus about the optimal treatment modalities for breast cancer in children is because of its absolutely rare prevalence. In this article, the medical history and treatment of a secretory breast carcinoma in an 11-year-old girl is reported. Modified radical mastectomy (MRM) was performed on April 6, 2013. Systemic chemotherapy was performed after surgery because metastatic lymph nodes were found in the dissected axillary tissue. Long term follow-up had to be done.

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Breast cancer in children represents less than 1% of all childhood malignancies [1] and less than 0.1% of breast tumors [2]. Levings published the first case of secretory (juvenile) carcinoma in a 12-year-old girl in 1917 [3].

Treatment of choice for patients with secretory breast carcinoma is controversial. Several authors suggest conservative surgery with sentinel lymph node biopsy [1,4,5]. Post-operative radiotherapy is applied in adult patients who have undergone quadrantectomy but this treatment is not advisable for children [4]. There are still no reliable data regarding the real therapeutic value of adjuvant chemotherapy.

1. Case report

A symptomatic 11-year-old girl was admitted to the hospital with a 3×3 cm lump in the subareolar region of her left breast. It had been detected 2 years before the operation and had had a slow growth pattern. A month prior to hospital admission history of trauma to the breast was negative. The mass was mobile and yellow nipple discharge was observed. No clinical axillary lymph node involvement was detected. A heterogenous mass with defined border was the result of son logic view, which was non-specific. Open surgical biopsy was performed. Microscopic examination revealed neoplastic tissue arranged in micro multicystic, tubuloalveolar and papillary patterns. The region was abundant with eosinophilic intracellular and extracellular secretory material (Fig. 1). The tumor cells were round to polygonal exhibiting moderate atypical the secretions in the lumen of micro cysts were PAS positive. These findings were compatible with secretory carcinoma.

On immunohistochemistry, the tumor cells were positive for S-100 protein but negative for estrogen and progesterone receptor and HER2.

^{*} Corresponding author. Department of Radiotherapy, Mashhad University of Medical Sciences, Vakilabad Street, Mashhad, Iran. Tel.: +98 (511)8525209. *E-mail addresses:* a.izanloo@yahoo.com, izanloua2@mums.ac.ir (M. Joudi).

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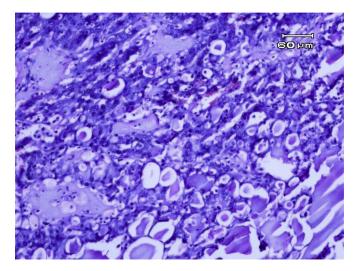


Fig. 1. Secretory breast carcinoma with micro multicystic pattern and abundant intracellular and extracellular secretory material (H & E, ×100).

Routine laboratory tests and serum prolactin were within the normal ranges. Modified radical mastectomy was performed on April 6, 2013.3 out of 12 resected lymph nodes were positive for metastatic carcinoma. Since this type of cancer is quite rare in children, and the literature review was limited to case reports, there is no specific medical law for the treatment planning at this age. Thus, we reviewed the case in a meeting with a radiotherapist, a chemotherapist, a pediatricians and a surgeon, discussing the treatment plan. Sentinel biopsy was overruled, because considering the removal of the breast tissues, there was no tissue left for sentinel biopsy. Chemotherapy was suggested for the patients after the surgery by prescribing cyclophosphamide, methotrexate and fluorouracil.

2. Discussion

Breast cancer is extremely rare among children; therefore, its clinical course is not even determined yet, resulting in disagreement on treatment planning. At First the description of the secretory breast carcinoma was in children, but later on it was detected among various age groups [6–8]. It usually occurs in females but was also reported among young males [9]. There is usually long time span between ... Long time span between the occurrence of symptoms and treatment irrespective of age and gender is usual [5]. In our case this period was 2 years of follow-up. Our 11-year-old patient had also positive familial history. Her aunt was treated for breast cancer, while BRCA1–2 mutations were negative. In the literature, a small number of patients have been reported to have appositive family history [1,6]. Positive familial history of breast cancer is a good indication of prognosis [6].

Despite its name, juvenile carcinoma, this distinctive pattern of carcinoma, is not limited to children and adolescents [10,4]. This tumor was named "secretory breast cancer" because tumor cells show abundant intra- and extracellular secretion and have granular

eosinophilic cytoplasm [4]. Secretory breast cancer is triple-negative (estrogen receptor-negative, progesterone receptor-negative, and HER2-negative), lacking the benefit of specific therapy that targets these proteins; nevertheless, it has a less aggressive behavior and a much better prognosis than the variants observed in adults do.

Axillary metastasis is rare and distant metastases are even rarer [1,5,7,8]. Local treatment should involve ablation or lumpectomy [1]. According to some authors mastectomy is the treatment of choice because of 75% local recurrence reports after BCS [6,7]. Sentinel lymph node biopsy is chosen over axillary dissection in order to avoid significant complications, especially in case of a child, who normally has a longer life span [1,10].

In general, in patients with small tumors and negative axillary metastasis, radiotherapy is not necessary after mastectomy [1]. Because of possible late side effects (lung and chest complications), radiotherapy must be avoided in children [10]. To decide on irradiation to morphological analysis of malignancy potential, including necrosis and vessel involvement, evaluation for Ki-67 expression and p53 over expression and hormone receptor status are necessary. Tumor size (less than 2 cm), age (less than 20 years at the time of diagnosis), and circumscribed margins (of the tumor) are favorable prognosis indicators [6].

3. Conclusion

Because hormone receptors are mostly absent; hormonal treatment should, therefore, be avoided [11]. Due to metastasis in the axilla, our patient received adjuvant chemotherapy, but at the present time there is not sufficient evidence in the literature to recommend adjuvant chemotherapy, even it has been pointed that no chemotherapy is warranted until the appearance of new lesions. Long term follow up should be done and a "wait-and-see" policy adopted.

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